



HIGH GRADE B CELL LYMPHOMA OF MANDIBLE - A CASE REPORT

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ABSTRACT

Non-Hodgkin's lymphomas are a group of neoplasms that originate from the cells of the lymphoreticular system. Non-Hodgkin's lymphomas detected primarily in the bone are quite rare, but among jaw lesions, they are more frequently present in maxilla than in the mandible. There are no classical characteristic clinical features of lymphomas involving the jaw bones. Owing to their low frequency, uncommon location and non-specific symptoms, Non Hodgkin lymphomas of the mandible are often misdiagnosed. We report a case of a large B-cell lymphoma of the mandible, in which the patient presented with a diffuse swelling on the right body of the mandible. A subsequent biopsy and immunohistochemistry were instrumental in the diagnosis.

KEYWORDS : Extranodal, Lymphoma, Non Hodgkin, Mandible

INTRODUCTION:

Lymphomas are malignant neoplasm of the lymphocyte cell lines. They are classified as either Hodgkin's or Non-Hodgkin's lymphoma (NHL). NHL comprises a heterogeneous group of lymphoid neoplasm with a spectrum of behavior ranging from relatively indolent to highly aggressive and potentially fatal. NHL presents up to 40% of the time at an extranodal site. Moreover, 2% to 3% of these extranodal cases may arise primarily in the oral cavity [1].

Oral lymphoma often is a component of a disseminated disease process that may involve regional nodes as well. Other times, it may represent as a primary extranodal disease confined to the oral cavity or jaws with mandible less frequently involved than maxilla as it accounts for only 0.6% of isolated malignant non-Hodgkin's lymphomas [2]. When it does occur, mandibular NHL typically manifests similar to an odontogenic process. This uncommon localization can pose significant diagnostic problems and is frequently misdiagnosed.

Case Report:

A 7-year-old male patient presented to the Department of Oral Medicine & Radiology with chief complaint of painless swelling in the right body of mandible since 3 years and growth on right side of face since 1 year. The swelling was insidious in onset and initially small in size, has gradually increased to attain present size over a period of 3 years without any associated symptoms. Medical history revealed that patient visited a private practitioner for swelling in the right parotid region and was operated for the same one year back following which growth appeared and attained present size within a duration of 1 year. Both dental and family history was noncontributory. A general physical examination revealed that patient was undernourished and poorly built.

Extra oral examination revealed gross facial asymmetry due to diffuse, smooth surfaced, non-erythematous swelling measuring approximately 4 X 3 cm on right side of mandible extending superiorly from 1cm above the lower border of mandible to 2 cm into the submandibular region. Mediolaterally the swelling extends from right outer canthus till angle of mandible. The surface of the swelling appeared stretched. There was no evidence of secondary changes over the swelling. On palpation, the swelling was non tender, uniformly firm in consistency, non - fluctuant, non - compressible, and was not adherent to the overlying skin. There was no paresthesia or lymphadenopathy associated with the swelling.

growth with definite and irregular borders on right side measuring approximately 8 X 4 cm in dimension. Superiorly it extends from level of right tragus of ear till the level of right angle of mouth, mediolaterally from right parasymphysis region to right ear lobe. Skin overlying the growth appears smooth, shiny and stretched with uniform contour and areas of hyperpigmentation were noted on its superior surface. There was no discharge or bleeding noted. The growth was firm, non-compressible on palpation and was fixed to underlying structures.

Intraoral examination revealed mixed dentition status with normal appearing mucosa. There was no evidence of draining fistulae, sinuses, or mobility of the teeth. However, #46 appeared to be lingually tilted. Based on history and clinical examination, we arrived at a provisional diagnosis of ameloblastoma of right body of mandible. Differential diagnosis of Non-Hodgkin's lymphoma, Ewing's sarcoma, osteomyelitis, and osteosarcoma were considered.

Panoramic radiograph shows a single, large, ill-defined, mixed radiolucent - radiopaque expansile lesion, extending from right body to ascending ramus of mandible and inferiorly approximately 2 cm below the lower border of mandible with destruction of the inferior border of mandible in the associated region. The internal structure of the lesion shows multiple radiopaque septa. CBCT showed massive destructive destruction involving right mandibular body and angle and ramus region with resorption of lower border in the concerned area.

An incisional biopsy of growth was performed that provided a diagnosis of keloid showing non keratinized stratified squamous epithelium and sub-epithelial connective tissue with basal layer showing presence of melanin pigment. The underlying connective tissue shows haphazardly arranged thick bundles of collagen giving glassy appearance with numerous fibroblasts and endothelial lined capillaries. Incisional biopsy of the swelling showed connective tissue stroma with sheets of intermediate and large round cells with open faced nuclei resembling lymphocytes. Nuclear polymorphism was also seen. On Immunohistochemical analysis, cells were LCA, CD20 and PAX5 positive, with Ki67 index of > 80 %. Reactive CD3 positive T cells were noted and BCL2 appeared focally positive. Hence, correlating with IHC, a final diagnosis of High grade B cell neoplasm was established.

Extra oral examination revealed solitary, sessile well defined

Extensive investigations including haematology, positron

emission tomography (PET) and CT were performed for head, neck, chest, abdomen and pelvis. The PET scan showed activity in the biopsy site of the right mandible. CT scan revealed no evidence of lymphoma in another site. The patient was then referred to a regional oncology center where prompt chemotherapy was instituted. Subsequently, the patient was examined periodically and a one-year follow-up evaluation confirmed remission.

DISCUSSION:

Lymphomas are a diverse group of neoplasms affecting the lymphoreticular system and are second only to squamous cell carcinoma in the frequency of malignant neoplasms involving the soft tissues of head and neck region, which usually affects the lymph nodes. Lymphomas have been divided into Hodgkin's disease and Non- Hodgkin's disease. Hodgkin's disease often presents as nodal disease, commonly involving cervical, axillary, and inguinal nodes, whereas non-Hodgkin's disease may develop extra-nodally, outside the lymphoid system and can occur in stomach, salivary glands, and rarely in oral cavity and jaws. NHL of bone is rare, representing only 5% of all extranodal lymphomas. They arise from the medullary cavity and manifest as a localized, solitary lesion [3].

It more commonly affects the middle aged and the elderly (40-80 yrs) with slight male predilection with a male to female ratio of 3:2 [3]. Primary lymphoma of the bone was first described by Parker and Jackson as primary reticular cell sarcoma of bone. Clinical features of lymphoma of the oral region are not characteristic. They occur as local bony swelling with associated tooth mobility, painless inflammation of the mucosa with or without ulcerations, and rarely as facial or dental pain. Additional observations include trismus, otalgia, gingival ulceration, sinusitis, or cervical lymphadenopathy[4]. The diagnosis of NHL of the mandible is frequently delayed or erroneously diagnosed as dental disease because the clinical presentation mimics that of an odontogenic process or localized osteomyelitis. There is an average lapse of 10-weeks between initial presentation and diagnosis [5].

There are no radiographic pathognomonic findings and are usually similar to those of periapical inflammatory process or osteitis. Diffuse trabecular honeycomb images are occasionally observed. Those may be the images of cortical destruction and invasion of the maxillary sinus. Features similar to that of non-specific osteolysis may also be present. Panoramic films usually show loss of cortical definition or widening of the mandibular canal and mental foramen, loss of lamina dura, or widening of periodontal ligament. Buccal or lingual cortical destruction is often found, but several reports note that NHL of the mandible can manifest as an extrasosseous soft tissue mass with only minimal cortical destruction observable on plain films. Differential diagnosis includes infectious process such as, dentoalveolar abscess, Wegener's granulomas, midline lethal granulomas, squamous cell carcinomas, metastatic tumors, neoplastic process, wherein very rapid growth is a feature of sarcomas and lymphoproliferative disorders [3].

Histopathologic evaluation, together with immunophenotypic and cytogenetic studies, elucidate the pattern of involvement and histologic type. When tissue diagnosis confirms NHL of the mandible, determination must be made regarding origination and spread of the tumor. The work up should assess the extent of disease and allow for accurate staging [6]. Following clinical examination and palpation of nodes, computed tomography is essential to exclude visceral or nodal involvement. A skeletal survey is usually not warranted without the presence of bone pain or related complaints. PET scanning is reported to be comparable to CT and provides no

additional information than the typical work-up. Laboratory studies are typically non-specific, although elevated lactate dehydrogenase is observed as a poor prognostic factor.

NHL can be managed by chemotherapy, radiotherapy, and surgery in various combinations. NHL arising in bone is best treated by chemotherapy and may not require radiotherapy [7]. Generally, a combination of chemotherapy (cyclophosphamide, doxorubicin, vincristine, and prednisone) and field radiation is recommended for treatment. Monoclonal antibodies directed against antigens or within the lymphoma and injection of interferon have also been used [8]. Survival is excellent in localized disease, whereas disseminated disease seems less favorable [7].

The prognosis of the disease is good with a maximum of 5-year survival rate in 30% of cases after therapy. The disease may occasionally progress into a diffuse pattern with cutaneous nodules and plaques which undergo blast transformation or rarely turn into leukemia [3].

We present a case of isolated mandibular NHL. This is a rare condition but one that should be considered in the differential diagnosis of swelling in the mandibular region. Fortunately, in our case, the solitary bony lymphoma had not disseminated and management by chemoradiation allowed for disease eradication.



1 (a) Lateral view of the extraoral swelling in the right mandibular region (b) Inferior view of the extraoral swelling in the right mandibular region



Figure 2 : Intraoral view showing lingually tilted #46



Figure 3: OPG showing mixed radiolucent –radiopaque lesion in right body and angle region of mandible

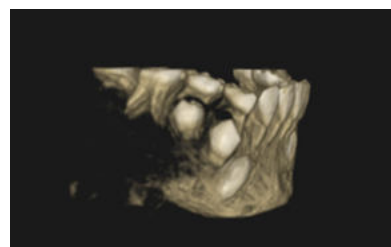


Figure 4: 3d Reconstructed Image Showing Massive Destruction In Right Mandibular Body And Angle Region With Complete Resorption Of Lower Border .

CONCLUSION:

The aim of this article is to develop awareness among dentists as there is an increased risk of oral NHL in the HIV-positive population and to emphasize the importance of including NHL in the differential diagnosis of intra oral soft tissues or ulceration to ensure appropriate treatment, to improve prognosis, and quality of life.

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